

## Case Report

# Anaesthetic management of giant encephalocele

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### ABSTRACT

One of the several challenges to the anaesthesiologists, is management of child with difficult airway. Management of even normal airway in a neonate is different and complex as compared to airway of two year old child and that of adult. Definition of the difficult airway is related solely to tracheal intubation or problems with mask ventilation. Among the different causes of difficult airway cranio facial and neoplastic anomalies are very common. We present a case report of difficult airway management in encephalocele patient.

**Keywords:** Encephalocele, Endotracheal intubation, Anaesthesia, Difficult airway

### INTRODUCTION

Neural tube defects constitute a major public health problem in term of mortality, morbidity, social cost and human suffering. Most infants are stillborn and live born infants survive for a short period of time, usually a few days.<sup>1,2</sup>

Encephaloceles are the result of a congenital cranial defect that allows intracranial contents to herniate. Primary arrest of bone development and adhesions from developing brain dura and skin further reduces bone development. Than increased intracranial pressure pushes the brain through the developing cranial base.<sup>3</sup> Incidence of encephaloceles is about 10 to 20% of all craniospinal dysraphisms.<sup>4</sup> Encephaloceles are classified by their anatomical location and their contents, Cranial meningoceles contains only leptomeninges and cerebro spinal fluid (CSF), whereas encephaloceles also contain brain, a ventriculocele is an encephalocele in which the herniated brain contents also contain portion of ventricle.<sup>5</sup> Occipital encephaloceles are usually more common (75%) than frontal (25%).<sup>6</sup>

There is no known genetic mutation present to explain the lesion, and higher incidence is in Southeast Asia (1:6000 live births), of that about 70% occur in females.<sup>7</sup> Prenatal diagnosis of encephaloceles is done by ultrasonography, maternal serum Alfa-fetoprotein (MSAFP) and amniocentesis.<sup>8,9</sup>

These lesions are mostly covered with normal, dysplastic skin or a thin, distorted meningeal membrane. The large encephalocele swellings may have brain herniation, abnormality of the underlying brain, microcephaly and ventriculomegaly, usually have poor prognosis.<sup>10</sup> Encephaloceles with dysfunctional brain tissue are treated by excision of the herniated brain tissue and repair of the defect in dura. The management of children with large dural defect along with herniation of a considerable proportion of brain matter is extremely difficult. In these cases preservation of the herniated brain parenchyma is done with expansile cranioplasty.<sup>11</sup>

## CASE REPORT

A male infant of 3 months old was admitted in neurosurgery outpatient department referred from paediatric department. The baby was born through lower segment caesarean section (LSCS) (Figure 1). There was a history of obstructed labor during birth. Baby was born after an uneventful 39 week pregnancy in a 27 year-old primipara. There is no record of prenatal diagnosis and baby cried immediately after birth. There is no history of any congenital anomaly in the family of mother and father, no history of consanguinity. There was a very big mass at the occipital region since birth, which was covered by normal pink-purple skin and hairs. Local examination revealed that swelling is consisting of three cysts like structure join together. Total circumference of the swelling is about 69 cm. The swelling was soft and transillumination test was positive (Figure 2), no bruit or murmur heard over the surface of the swelling. Anterior and posterior frontanelle were opened.



**Figure 1: Infant at birth.**



**Figure 2: Transillumination test.**

The infant used to lie in lateral position, because of the swelling. Neck remains in flexion and extension is not possible in supine position. No facial, maxillary and mandibular defects were present. No other congenital anomaly was present. Weight of infant was 5.9 kg.

On neurological examination, the baby was conscious with no limb weakness and accepting breast feeding normally. There was Pupils were normal and reacting to light.

Magnetic resonance imaging (MRI) of brain showed a giant encephalocele at the occipital region (Figure 3),

other investigation was within normal limit, and no other congenital anomaly was detected in the infant.



**Figure 3: MRI.**

Infant was posted for excision of encephalocele sac under general anesthesia. A 4 hour fasting was done before operation.

Operation theater (OT) was prepared before shifting the infant, Devices for maintaining normothermia like circulating warm-air devices (Bair Hugger paediatric 530), airway humidifiers, and fluid warming devices (Belmont Buddy Lite) should be kept ready. An alternative airway management plan also be kept ready that includes laryngeal mask airway of appropriate size, with high frequency jet ventilation, fiberoptic bronchoscope, a cricothyroid cannula and preparations for tracheostomy, after taking infant inside the OT a warming blanket was placed over the baby. ECG, NIBP, SPO2, ETCO2, and Precordial stethoscope was also attached for monitoring. Baseline parameters of the baby were recorded. Intra venous fluid isolyte P 15 ml/hour was started. The baby received inj. glycopyrrolate 0.03 mg intravenously (IV) and inj. fentanyl 10 µg as premedication. Baby was induced in supine position with inj. propofol 15 mg and inj. succinylcholine 15 mg was given after confirming adequate mask ventilation in lateral position, using 100% oxygen. Intubation was done by lifting the baby and placed her head beyond the edge of table with an assistant supporting it while another assistant stabilized the baby's body, taking adequate care to support the encephalocele so as to prevent a rupture. Baby was intubated using 4.0 mm inner diameter uncuffed endotracheal tube and was fixed at 12 cm after confirming bilateral equal air entry. After proper tube fixation, the baby was made prone with extreme care to prevent accidental extubation (Figure 4). Anaesthesia was maintained using oxygen, nitrous oxide (50 : 50), halothane (0.5%) and inj. atracurium 5 mg bolus and 1mg top ups. Paracetamol suppository 100 mg was inserted per rectally. At the end of the surgery, N<sub>2</sub>O discontinued and patient was reversed using inj. Neostigmin 0.25 mg and inj Atropine 0.2 mg/kg when spontaneous respiratory efforts were noticed. The patient was extubated when adequate spontaneous respiratory efforts were returned and child became conscious. The child started crying after extubation and was shifted to PICU with O<sub>2</sub> for

further monitoring. Post operatively, child was observed for respiratory complications.



**Figure 4: After intubation.**

## DISCUSSION

While caring for infant with occipital encephalocele our aim were to avoid premature rupture of the encephalocele and to manage a possible difficult airway due to restricted neck movement and inability to achieve optimal position for endotracheal intubation.

Airway management in pediatric patients with poses many challenges to the anesthesiologist. Isada et al. reviewed 13,557 pediatric cases of craniofacial malformation and reported that the risk of difficult intubation is higher in children with congenital malformation.<sup>12</sup> Another study on 118 children of encephalocele shows difficulty during endotracheal intubation in 19.5% cases.<sup>13</sup>

Airway management of occipital encephalocele children were explained by various authors. Quezado et al described a foam-cushion device to prevent pressure on encephalocele sac.<sup>14</sup>

Quezado et al<sup>14</sup> and Mowafi et al<sup>15</sup> have put blanket below baby trunk with the head hanging from the edge of the table to facilitate intubation in such patients, while Manhas et al took aid to lift the child (one person to support the head and shoulder and a second person to lift the torso and legs) for intubation after attempts to intubate in the lateral position failed.<sup>16</sup> Dey et al used a third person to extend the neck to successfully intubate a neonate in whom the two earlier described techniques at intubation failed.<sup>17</sup>

Other aids for securing airway in newborn and infants are now available like Laryngeal mask airway, Video Laryngoscope are being used to facilitate intubation in neonates and smaller children in difficult situations. But use of these devices requires familiarity and experience before they can be successfully used.

Other concerns that need the anesthesiologist's attention in patients with occipital encephalocele are raised

intracranial pressures, low cerebral perfusion pressures, disturbances in central autonomic control and temperature regulation in these children.

In the presence of an encephalocele, there is a 60-80% risk of associated structural abnormality both intra-and extra cranially in patients. Postoperatively patients may also develop hydrocephalus requiring a ventriculoperitoneal shunt.<sup>18,19</sup> Survival rates and morbidity of encephalocoels vary most strongly with anatomical sites being 100 and 50% respectively in the case of anterior defects and 55 and 83% respectively in the case of posterior defects.<sup>20</sup>

Patients with giant encephalocele and large amount of brain tissue in the sac usually die either shortly after birth or as a result of operation. A microcephalic child with neurological deficit and a sac containing cerebrum, cerebellum and brain stem structures, carry a poor prognosis. In such patients, it is generally impossible to foretell whether the infant will die quickly or will continue to live for many months or years, as size of the encephalocele itself is not a guide to prognosis. Ultimate result depends on the amount of normal brain tissue left inside the skull after the operation.<sup>20</sup>

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